Retinitis Pigmentosa: a review and update on treatment and management.

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A 26-year-old Hispanic male presents to confirm the diagnosis of retinitis pigmentosa (RP) and explore possible management options. Reverse telescopes and prism can improve orientation and assist with mobility. Current research will be discussed.

I. Case History

- **Demographics:** 26-year-old Hispanic male
- **Chief complaint:** Gradual peripheral field loss since 8th grade, increasing difficulty with mobility at night and adjusting from dark to bright environments
- **Goal:** Improve mobility, driving, return to employment
- **Ocular history:** Unremarkable
- **Medical history:** Patient reported wearing hearing aids since age three. Family history includes a younger sister who has worn hearing aids since age one.
- **Medication:** None, NKDA
- **Occupation:** Recently unemployed, was working as a radiology technician

II. Pertinent findings

- **Visual Acuity:** BCVA 20/30+ OD and OS
- **Visual Fields:**
  - HVF 30-2: constricted to 15 degrees OD, constricted to 20 degrees OS
  - Binocular Esterman: constricted to 35 degrees
- **Fundus exam:** Minimal vessel attenuation, no signs of bone-spicule pigmentation or waxy optic nerve pallor
- **Special Testing:**
  - ERG showed diminished a- and b-wave
  - OCT of the retina showed absence of photoreceptor layer throughout the posterior pole
  - Dynavision showed decrease in reaction time to peripheral stimulus
  - Useful Field of View revealed a low to moderate risk (category 3) of collision while driving
- **Devices:** Patient showed good positive response to demonstration of 2.8x and 4x12 reverse hand-held telescope along with sector prism for field enhancement.

III. Differential diagnosis

- **Primary Dx:** Retinitis pigmentosa
- **Others:** Usher’s syndrome, cone-rod dystrophy, congenital stationary night blindness

IV. Diagnosis and discussion
Although there were no clinical signs of bone-spicule pigmentation, the results of ERG, OCT and visual field testing confirmed the diagnosis of retinitis pigmentosa. The diagnosis of Usher’s syndrome is inconclusive at this time pending further testing.

V. Treatment, management

- Treatment and response to treatment
  - The patient was educated on the impact of retinitis pigmentosa on visual function.
  - The patient was advised against driving based on results of visual field testing, Dynavision and Useful Field of View in accordance to Texas Department of Public Safety Medical Advisory Board recommendation of 140 continuous degrees of visual field.
  - The patient was educated on alternate transportation options as well as local support groups.
  - A referral for orientation and mobility training was discussed to assist with daily activities, especially at night or dimly-lit environments.
  - The patient responded well to visual field enhancement using a hand-held reverse telescope and sector prism on his glasses. The patient chose to explore these options further at a later time.

- Referral for research as appropriate
  - There are currently no treatment options for retinitis pigmentosa other than low vision rehabilitation.
  - The patient was educated on current research using valproic acid and vitamin supplementation to reduce the progression of visual field loss.
  - The Argus® II Retinal Prosthesis System can provide an improvement in visual perception for daily activities in patients with retinitis pigmentosa with profound vision loss.

VI. Conclusion

- RP is a devastating disease that not only slowly diminishes a patient’s visual field but also can disrupt their normal daily activities. It is imperative that the physician approach this holistically, with reassurance and sensitivity, while providing the resources and assistance to maintain the patient’s independence and quality of life.

References


